Microvascular Right-to-Left Pulmonary Shunt Demonstrated by a Radionuclide Method

Erik Kjøller, Flemming Pedersen, Tage Lysbo Svendsen, and Sven Sølvsten Sørensen

Cardiovascular Laboratory and Department of Clinical Physiology and Nuclear Medicine, Rigshospitalet, University Hospital, Copenhagen, Denmark

A 37-yr-old man with angiolymphoid hyperplasia (Kimura's syndrome), who had been treated unsuccessfully for suspected asthma, was investigated due to a decrease in arterial oxygen saturation (86%). Right heart catheterization and angiography of the pulmonary artery failed to demonstrate any right-to-left shunts. However, simultaneous scintigraphy over the lungs, kidneys, and head after injection of 150 MBq technetium-99m-labeled macroaggregated albumin i.v. and inhalation of 150 MBq krypton-81m demonstrated a right-to-left shunt in the lungs probably caused by precapillary pulmonary arteriovenous shunts.

J Nucl Med 1991; 32:139–140

Right-to-left shunts are generally diagnosed by right heart catheterization including angiography. If this investigation fails to confirm the diagnosis but the clinical suspicion persists, other investigations might have to be performed. One of the possibilities is dynamic whole-body scintigraphy, which, in the presence of intrathoracic right-to-left shunts, has been able to confirm the diagnosis (*1–3*). In the case presented, this method was the only way to make a probable assessment on the presence of a pulmonary right-to left-shunt.

CASE HISTORY

A 37-yr-old man with known angiolymphoid hyperplasia with eosinophilia (ALHE) (Kimura's disease) (*4*) presented with increasing dyspnea and a decrease in arterial oxygen saturation. He had been well until 1971, when he reported pain in the right knee. The following year he observed several brown-blue nodules around the knee. On hospital admission in 1978, a 4 × 4 cm pulsating swelling was perceptible with a rumbling murmur heard by auscultation. Angiography showed an angiomatous structure anterior to the popliteal vessels containing multiple, tortuous arteriovenous communications with several sacculated aneurysms. The aneurysms and subcutaneous swellings were resected and histology showed changes consistent with ALHE. During the following years, several subcutaneous nodules were resected from all over the body—in each case the histology was consistent with ALHE. In 1982, the patient was operated on for a brain abscess due to *staphylococcus aureus*. The surgical intervention was uncomplicated and the patient was without sequelae. No histology was obtained.

Since 1979, the patient had suffered from dyspnea at exercise. Based on clinical judgment and the presence of eosinophilia in the blood and a slight increase of immunoglobulin E, the patient was believed to be suffering from asthma bronchiale. He was treated with conventional asthma therapy without great success. All tests to confirm allergic reactions were negative. In 1987, a decreased arterial oxygen saturation was found and he was transferred to our department for further investigations.

Physical examinations showed a slight cyanosis of the lips and fingers and pronounced clubbing of the fingers. Several red-brown-blue nodules measuring up to 2.0 cm in diameter were noted around the body. The lungs were clear. Auscultation of the heart as well as abdominal examination was normal. There was no peripheral edema. Hemoglobin was 9 mmol/l with normal hematocrit. White cell counts were 7.3 × 10^9/l with increased eosinophils 11%. The platelet count was 268 × 10^9/l. Serum-sodium, potassium, calcium, and creatinine were all normal, as were the liver function tests. Plasma fibrinogen, -alpha-1-antitrypsin, and immunoglobulin G, M, and A were all normal, whereas immunoglobulin E was increased.

Oxygen saturation in arterial blood had decreased to 86%, pO_2, 6.7 kPa and pCO_2, 5.65 kPa. All values were unchanged during maximal symptom-limited exercise. Lung function tests were normal (FEV1: 3.5 liter/sec, vital capacity 4.9 liters and residual volume 1.75 liters). Oxygen diffusion capacity measured by carbon monoxide was normal: 8.37 mmol/l/kPa (8.08–14.39)/l alveolar volume 1.40 ml/min kPa (1.20–2.29). ECG was normal as were X-ray and CT scans of thorax. Two-dimensional and M-mode echocardiography was normal. A right heart catheterization showed normal pressures. There were no signs of intracardiac left-to-right shunt: mean vena cava oxygen saturation 77% and pulmonary arterial oxygen saturation 78%. A selective right and left pulmonary arteriography was without any evidence of arteriovenous shunts.

Since ventilation/perfusion mismatch was suspected, a pulmonary scintigraphy (General Electric Maxicamera 535) was performed after the i.v. injection of 150 MBq technetium-99m-labeled macroaggregated albumin (^{99m}Tc-MAA) and the inhalation of 150 MBq krypton-81m (^{81m}Kr). There was an even distribution of activity on both ventilation and perfusion scintigrams. Hence, there was no evidence of pulmonary

Received Jun. 13, 1990; revision accepted Jul. 6, 1990.

For reprints contact: Dr. Erik Kjøller, Medical Department F, Herlev Hospital, University of Copenhagen, DK-2730 Herlev, Denmark.

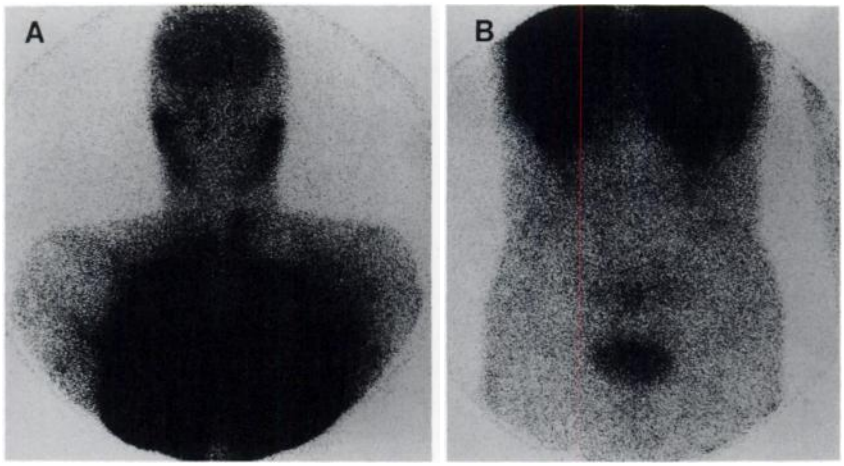

FIGURE 1
Scintigrams following i.v. injection of 200 MBq ^{99m} Tc-labeled MAA. There is activity uptake in lungs, cerebrum, thyroid, and salivary glands (A) and urinary bladder (B).

emboli. However, activity was seen in the kidneys and in the face suggesting the presence of pulmonary arteriovenous communications.

Later scintigraphy was performed simultaneously over the lungs and kidneys following i.v. injection of 200 MBq ^{99m} Tc-labeled MAA. Scans were obtained dynamically over 11 min. Subsequent static pictures were obtained in two regions including: (a) lungs and kidneys and (b) lungs, neck, and head regions to estimate the fractional distribution of cardiac output (*5*). The dynamic pictures revealed a rapid increase of activity over both lungs and kidneys within the first minute, reaching its maximum after 1 min. In the static images, activity was seen (apart from lungs and kidneys) in the thyroid gland (Fig. 1A-B), parotics, and in the cerebrum. Based on the count rate in lungs and kidneys and given a kidney flow of 20%–25% of cardiac output an approximate right-to-left shunt of 10%–20% could be calculated. The free activity in the injected preparation was less than 0.5%. It was concluded that the investigation was consistent with a right-to-left shunt at the pulmonary level.

DISCUSSION

The permanent decrease of arterial oxygen saturation raised the suspicion of intracardiac right-to-left shunt, pulmonary arteriovenous fistulae, or primary pulmonary disease. Intracardiac shunt was ruled out by the normal heart catheterization. The presence of shunts after injection in both left and right arm precluded extra cardiac venoarterial shunts due to anomalous systemic venous drainage (*2,3*). Patients with pulmonary arteriovenous fistulae have changes on X-ray of the chest and the shunts are easily demonstrable on pulmonary angiography (*6*). A right-to-left shunt could still not be excluded due to the prior cerebral abscess (*7*). Primary pulmonary disease was ruled out by normal lung function tests. Asthma was ruled out by the lack of effect of treatment including steroids. Eosinophilia (*8*) and increased IgE are seen in ALHE and thus cannot support the diagnosis of asthma bronchiale. Asthma has previously been suspected in ALHE but no thorough examination has been performed in these patients (*9*).

The early appearance of systemic activity after i.v. injection of radioisotope demonstrated the presence of intrathoracic vascular right-to-left shunt, as was the case in previous studies (*1,2,3*). This early appearance could also be due to a high amount of free activity in the injected preparation. The measured free activity was low, however, and thus could not explain the sudden appearance of high activity in the systemic circulation.

A precapillary pulmonary arteriovenous shunt was suspected because the patient previously had demonstrated arteriovenous fistulae (*3*) due to ALHE. Although ALHE has been demonstrated in the lungs, the possible clinical effect of an arteriovenous shunt has never been demonstrated.

REFERENCES

1. Konstam MA, Levine BW, Strauss, HW, McKusick KA. Left superior vena cava to left atrial communication diagnosed with radionuclide angiography and with differential right-to-left shunting. *Am J Cardiol* 1979;43:149–153.
2. Verzijlbergen F, Telling C, Van Plokker MWM. Significance of the site of injection in unexpected right-to-left shunting. *J Nucl Med* 1984;25:1103–1105.
3. Rosenbaum RC, Reiner BI, Bidwell JK, Johnston GS. Right-to-left shunting via persistent left superior vena cava identified by perfusion lung scintigraphy. *J Nucl Med* 1989;30:412–414.
4. Moesner J, Pallesen R, Sørensen B. Angiolymphoid hyperplasia with eosinophilia (Kimura's disease). *Arch Dermatol* 1981;117:650–653.
5. Crean PA, Pratt T, Davies GJ, Myers M, Lavender P, Maseri A. The fractional distribution of the cardiac output in man using microspheres labelled with technetium-99m. *Br J Radiol* 1986;59:209–215.
6. Moyer JH, Glantz L, Brest AN. Pulmonary arteriovenous fistulas. *Am J Med* 1962;32:417–435.
7. Brendel AJ, Larnaudie B, Lambert F, et al. Unsuccessful lung scan due to major right-to-left shunt through a sinus venous septal defect. *J Nucl Med* 1985;26:1029–1034.
8. Olsen TG, Helvig EB. Angiolymphoid hyperplasia with eosinophilia. *J Am Acad Dermatol* 1985;12:781–796.
9. Saxe N, Kahn LB. Angiolymphoid hyperplasia with eosinophilia. *S Afr Med J* 1977;52:454–457.